

Topics: Very Young Children; Tethered Cord; Related Conditions; Morphometrics

Conquer Chiari's monthly research updates highlight and summarize interesting publications from the medical literature while providing background and context. The summaries do contain some medical terminology and assume a general understanding of Chiari. Introductory information about Chiari, plus many more research articles, can be found at www.conquerchiari.org.



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 A surgical team from Northwestern University in Chicago reported on their experiences in surgically treating very young children with Chiari. According to a national pediatric database, most Chiari cases in children occur between the ages of 5-15; however, symptomatic Chiari is also seen, less frequently, in infants and toddlers. Specifically, these clinicians surgically treated 16 children under the age of 3 over a 7-year span, and followed them for a minimum of 3 years. Symptomatically, 75% (12) showed pre-verbal signs of headache, such as inconsolable crying, head grabbing, and back arching. Throat and airway issues were also very common, with 10 children experiencing symptoms such as vomiting, choking, gagging, sleep apnea, and vocal cord paralysis. MRIs showed the average tonsillar descent was a sizable 9.8mm. Two of the children had hydrocephalus, but only one had syringomyelia. Unfortunately, the initial surgical success rate was not that great. While all the young patients showed initial improvement, nearly half (7) had symptoms come back and required a second surgery. Interestingly, whether the dura was fully opened during the first operation was not statistically linked to whether additional surgery was required.

SOURCE: *Chiari type I malformation of infants and toddlers. Grahovac G, Pundy T, Tomita T. Childs Nerv Syst. 2018 Feb 2.*

 Some in the Chiari community have speculated that there is a strong, even causative link between tethered cord and Chiari malformation. In other words, they theorize that the pulling traction at the bottom of the cord that exists in tethered cord is translated up and somehow results in the cerebellar herniation that defines Chiari. While there is currently little evidence to support this theory, an interesting case study from doctors at Harvard and Johns Hopkins actually goes the other way. They report on an adult patient who developed tethered cord after Chiari decompression surgery. This fascinating case focuses on a 68-year-old man who had a traumatic fall more than 30 years earlier. After lower spinal surgery, he developed post-traumatic syringomyelia which was treated with a shunt placed directly in the syrinx. He was neurologically stable for decades but then late in life developed symptoms common to Chiari. An MRI indeed showed tonsillar herniation and a cervical syrinx, so he underwent decompression surgery. About two weeks after the surgery, his legs became weak to the point he could no longer walk. Imaging of the lower spine showed clear indications of cord tethering near his old surgical site with complete CSF blockage. Subsequent spinal surgery improved his leg strength somewhat but not completely. While cord tethering after a fall and spinal surgery would not be considered unusual, it is not at all clear why it occurred in this case decades later, with symptoms apparently sparked by Chiari decompression surgery. The authors speculate that the altered CSF dynamics from the Chiari surgery somehow played a role.

SOURCE: *Adult tethered cord syndrome following Chiari decompression. Jackson C, Yang BW, Bi WL, Chiocca EA, Groff MW. World Neurosurg. 2018 Jan 31.*

 [Note: The author, R Labuda, is also a listed author on the publication discussed below] Researchers from the Conquer Chiari Research Center continued to leverage the success of the Chiari 1000 platform with a recent publication which used morphometrics to compare Chiari patients with conditions commonly related to Chiari. Using images shared through the Chiari 1000, the CCRC grouped 236 adult women as either having, or not having, one of five commonly related conditions such as pseudotumor, EDS, and syringomyelia; and then also 7 highly prevalent conditions, such as chronic fatigue, fibromyalgia, and migraines. For each of the 12 conditions, the researchers used 28 morphometric measures to compare the Chiari patients who had a specific additional condition with those who didn't. They also then compared all the patients with a specific additional condition to a large group of healthy controls. Among the additional conditions, they did not find as many differences as they were expecting, but there were a couple. Specifically, patients who also syringomyelia had significantly narrower foramen magnums, about 2mm less, than those patients without syringomyelia. Also, on average, the tonsillar descent of patients with EDS was not as large as those without EDS (5.54mm vs 8.37mm respectively). In comparing the related condition groups to the healthy controls, the researchers found there were a number of morphometric differences for each related condition. However, interestingly, four of these measures were consistently different across all of the related condition groups. For example, every group showed a significantly shorter clivus bone than the healthy controls. The clivus bone has been noted previously as potentially playing a key role in the development of Chiari anatomy, so this is a potentially important finding. This study was published in an open access journal and the full text of the article can be found here:

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5785719/>

SOURCE: *A Retrospective 2D Morphometric Analysis of Adult Female Chiari Type I Patients with Commonly Reported and Related Conditions. Eppelheimer MS, Houston JR, Bapuraj JR, Labuda R, Loth DM, Braun AM, Allen NJ, Heidari Pahlavian S, Biswas D, Urbizu A, Martin BA, Maher CO, Allen PA, Loth F. Front Neuroanat. 2018 Jan 19;12:2.*

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